BPA Questionnaire on Acute porphyrias

data collected Spring/Summer 2006

There is surprisingly little known about where and how those with acute porphyria are actually treated. We therefore sent out this questionnaire to those on our books with acute porphyrias, to gather information on that, and related questions.

We deliberately did not make the questionnaire too detailed, so it would not take too long to complete.

We limited the questionnaire to those with acute porphyrias – AIP, VP, HCP (Acute Intermittent Porphyria, Variegate Porphyria and Hereditary Coproporphyria)

So **not** those with EPP, CP or PCT (Erythropoietic Protoporphyria, Congenital Erythropoietic Porphyria or Porphyria Cutanea Tarda)

We sent out 185 questionnaires	**92 were returned**:	50%
109 Were to paid-up members:	71 returned:	65%
77 were to non-members	21 returned:	27%

So there was a good response from the paid-up members of BPA.

Not all respondents filled in all the questions, so numbers for specific answers vary.

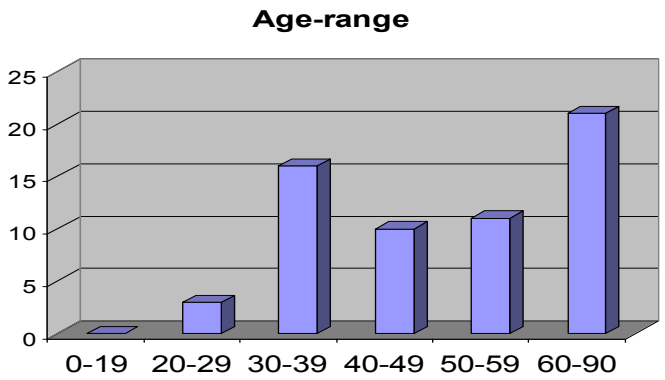
Throughout this report, I have used normal statistical tests to check if differences are meaningful, or due to chance and the low numbers (I used the normal 95% statistical confidence).

Information on respondents:

Age

61 respondents gave their age

0-19	0
20-29	3
30-39	16
40-49	10
50-59	11
over 60	21
total	**61**



The great majority were over 30.

Gender

women	74	80%
men	18	20%
total	**92**	

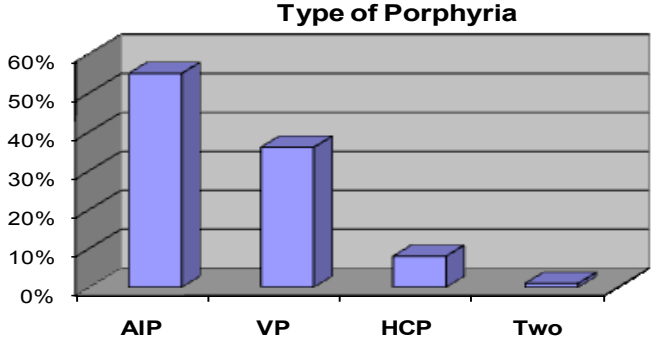
	female	male	total
suffered attacks	69 (88%)	9 (12%)	78 (100%)
symptom free	5 (36%)	9 (64%)	14 (100%)

The symptom free respondents had DNA-confirmed porphyria, after a relative showed symptoms

The split between sexes for them is not significantly different to 50/50.

Type of Porphyria

AIP	51	55%
VP	33	36%
HCP	7	8%
Two of these	1	1%
total	**92**	

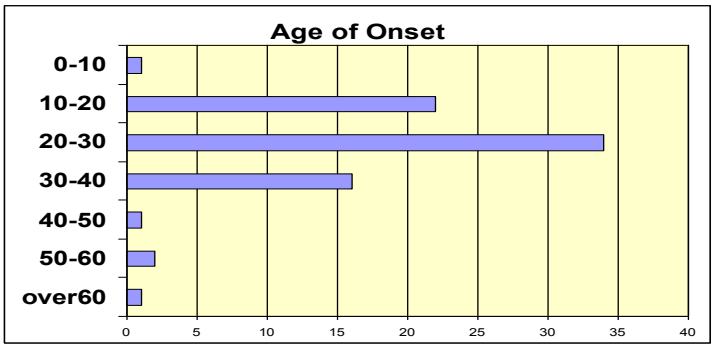


all types had 80% female / 20% male (within statistical variation)

The split between types of porphyria, within the respondents, is similar to the split within the total BPA membership and also the split within the general population.

Age of On-set

Age	Number
0-10	1
10-20	22
20-30	34
30-40	16
40-50	1
50-60	2
over60	1
Total	**77**

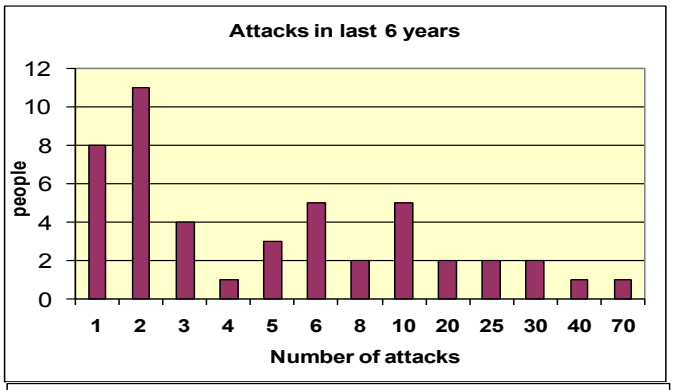


The great majority started with attacks of porphyria in their teens, twenties or thirties.
Note that many of the respondents are now much older.

Number of attacks in last 6 years

No. of attacks	people
1	8
2	11
3	4
4	1
5	3
6	5
8	2
10	5
20	2
25	2
30	2
40	1
70	1
lots	1

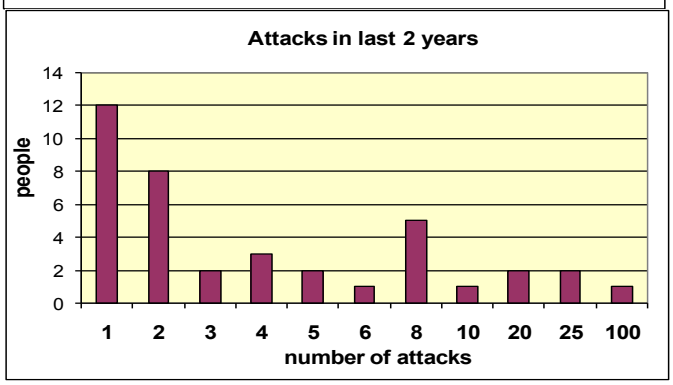
Total 48

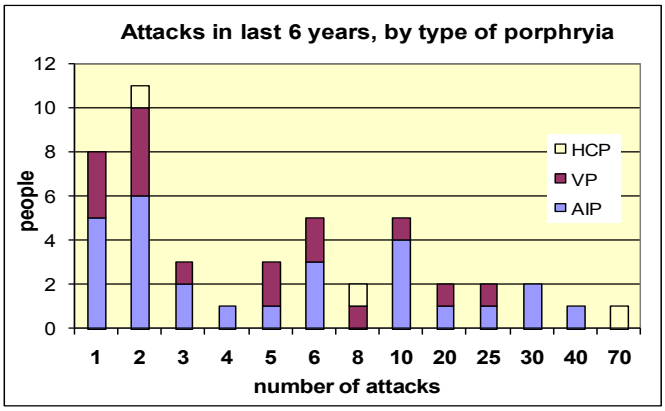


Attacks in last 2 years

No. of attacks	people
1	12
2	8
3	2
4	3
5	2
6	1
8	5
10	1
20	2
25	2
100	1

Total 39





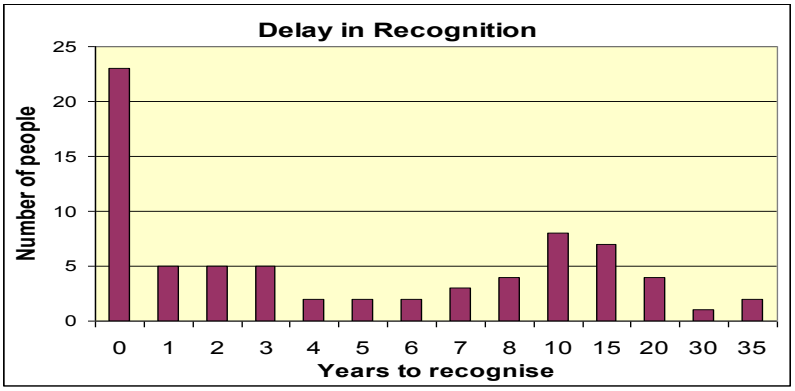
"people" means number of respondents

This shows half having one or a few attacks (up to 3 in 6 years), and half having more.
These range from one attack a year, to one a month or more.
(It is a classic double-humped distribution)
The number of attacks does not depend on the type of acute porphyria.

Delay in identifying porphyria

These four graphs show the delay between having the first symptoms of porphyria, and when it was recognised that porphyria was the cause of these symptoms.

Years	People
0	23
1	5
2	5
3	5
4	2
5	2
6	2
7	3
8	4
10	8
15	7
20	4
30	1
35	2
Total	**73**



So a third are recognised within a year,
just over half are recognised within 3 years but a third take 10 years or more to be recognised – up to 35 years.

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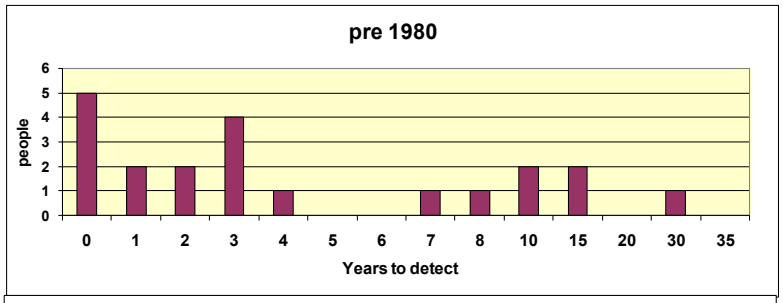
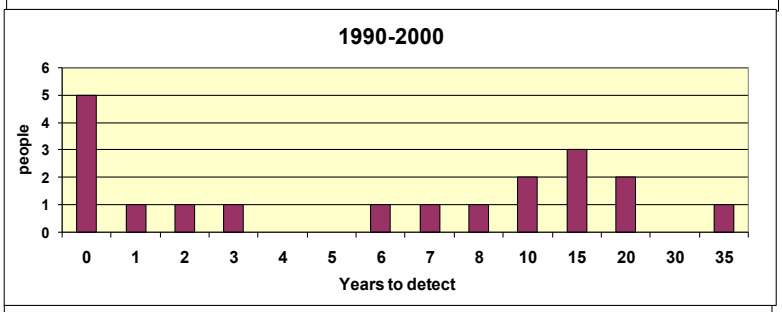
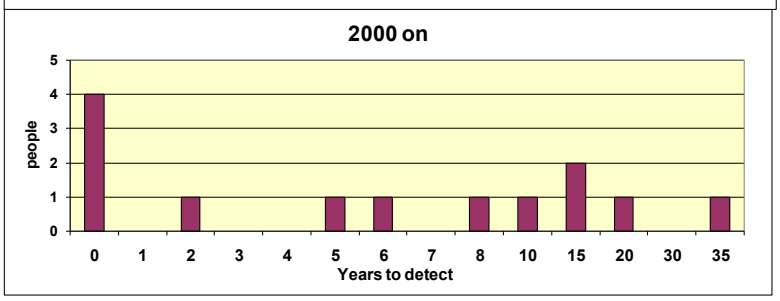
In these three graphs, they are grouped by the decade in which porphyria was recognised.



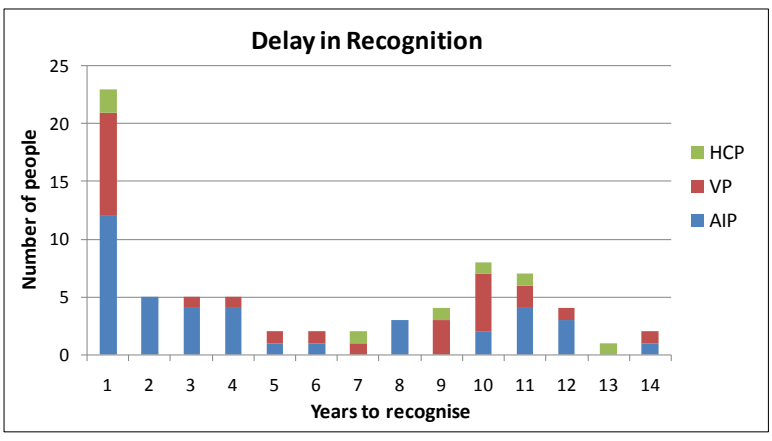


There has not been any significant improvement in the speed of recognition of porphyria over the last few decades.



Types of porphyria:

There are no significant differences in delay of recognition between the different types of porphyria.



Hospitals

The question on "**which hospital diagnosed porphyria**?" came up with a total of **64** different hospitals.

Those occurring 3 or more times were:

Hospital	Location	patients
UHW	Cardiff	13
Hallamshire Hospital	Sheffield	4
Kings College	London	4
UCH	London	3
Kent & Sx Hospital	Tun. Wells	3
Southend Hospital	Southend	3
(Others		34)

Cardiff came top, because of its central role of genetic and biochemical testing. But most patients were diagnosed in their own local hospital.

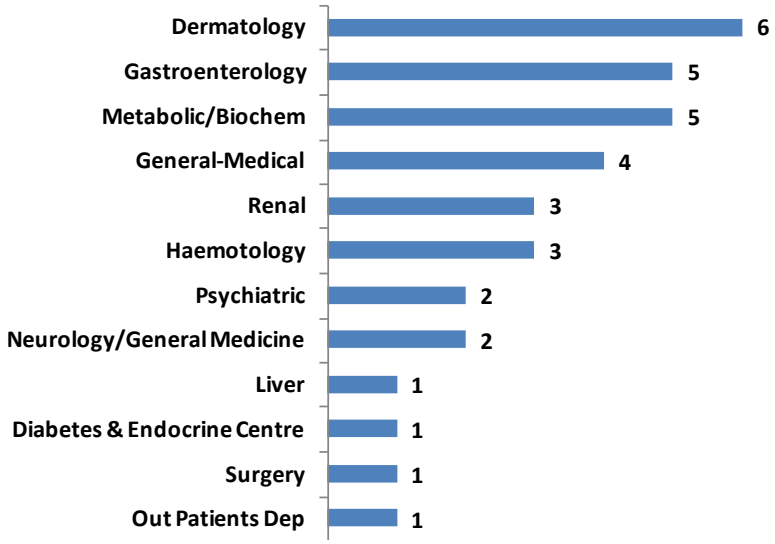
The question on "**which hospital is currently treating you?**" was answered by 56 people, and came up with a total of **42** different hospitals.

Those occurring 2 or more times were:

Hospital	Location	patients
Kings College	London	6
Addenbrookes	Cambridge	4
UHW	Cardiff	2
Q Elizabeth	Birmingham	2
Derby City General	Derby	2
St James's	Leeds	2
Bassetlaw Hospital	Worksop	2
MRI	Manchester	2
(Others		26)

Again, other than Kings and Addenbrookes, there is no concentration of treatment of porphyria in hospitals specialising in the disorder.

Specialism of consultant dealing with their porphyria



There was an amazing range of specialisms dealing with porphyria patients, with gastroenterology near the top!

Occurrence and "Severity" of attacks

One of the most significant outcomes of the questionnaire was on the replies to the questions on the occurrences of attacks.

Of the 92 respondents:

14	(15%)	were symptom-free (had a relative diagnosed)
29	(32%)	had attacks over 6 yrs ago
10	(11%)	had attacks in the last 6 years, but not the last 2 years
39	(42%)	had attacks in the last 2 years

Of the 39 who had had attacks in the last 2 years:

Only	16	of them had been severe enough to need treatment in hospital
So	23	treated themselves at home

So, in summary:

1: symptom free	14	(diagnosed after a relative had porphyria)
2: attack over 6 yrs ago	29	
3: attack 3 to 6 yrs ago	10	
4: attack but not hospitalised	23	
5: hospitalised	16	
Total	**92**	

A few of the 23 who treated themselves at home put "mild attacks only" on the form, but most just entered the number of attacks.
Also another 3 said they had medical problems, but did not know if they were due to their porphyria.

So this questionnaire makes it clear that porphyria has a much bigger effect on the respondents' lives than is given by figures of severe attacks seen by hospitals.

Two typical comments made on the questionnaires:

"I have not had a very bad attack since 1999.
However I have quite a few illnesses and symptoms which I believe are from Porphyria. Weakness is a constant one, and I find they are not often viewed as part of Porphyria symptoms. Also I can get quite down, and feel very shaky." (AIP)

"My problems are mainly to do with my skin.
When I was diagnosed I was told it 'won't affect your every day life'. How wrong was that comment. I have to take care doing housework, walking through doors carrying things, reaching for items in confined spaces, not to mention trips to the doctors." (VP)

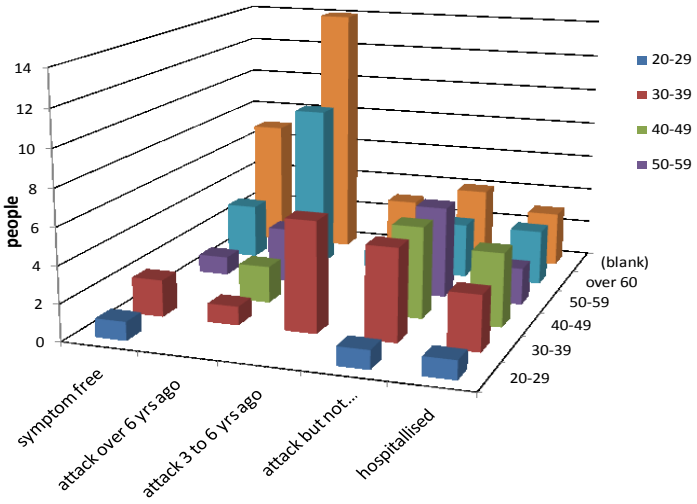
Severity and age range

	20-29	30-39	40-49	50-59	over 60	**Total**
symptom free	1	2	-	1	3	7 people
attack over 6 yrs ago	1	2	3	9	-	15 people
attack 3 to 6 yrs ago	-	6	-	-	1	7 people
attack but not hospitalised	1	5	5	5	3	19 people
hospitalised	1	3	4	2	3	13 people
Total	**3**	**17**	**11**	**11**	**19**	**61 people**

Those with attacks 3 to 6 years ago, but none in the last two years are mainly in their 30s.

Those who had attacks over 6 years ago, tend to be older.

The other groups are all split between ages in similar ways to each other.



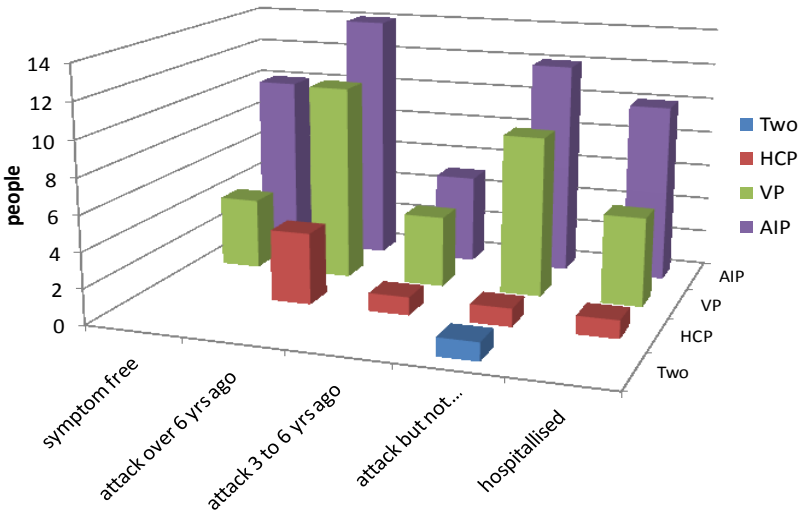
Severity and type of porphyria

	Type of porphyria				
	AIP	VP	HCP	Two	Total
symptom free	10	4	-	-	14 people
attack over 6 yrs ago	14	11	4	-	29 people
attack 3 to 6 yrs ago	5	4	1	-	10 people
attack but not hospitalised	12	9	1	1	23 people
hospitalised	10	5	1	-	16 people
Total	**51**	**33**	**7**	**1**	**92 people**

Those with HCP have mainly been free of attacks in the last 6 years.

The other groups are all split between types of porphyria in similar ways to each other.

Numbers are different because different respondents answered different questions.



Severity and gender

	total	female	male	% f	% m
symptom free	14	5	9	36%	64%
attack over 6 yrs ago	29	27	2		
attack 3 to 6 yrs ago	10	9	1		
attack but not hospital	23	19	4	88%	12%
hospitalised	16	14	2		
Total92	74	18			

The proportion of males and females without symptoms does not differ significantly from 50 / 50.

None of the other groups differ significantly from the overall 88% female.

Treatment

The 16 people hospitalised in the last 2 years were treated as follows:

Haem arginate and Glucose	6 people
Haem arginate only	0 people
Glucose only	9 people
Nothing	1 person

Glucose only: All were in different hospitals.

Haem arginate: 3 given without albumen, 1 with albumen, 2 did not know.

One respondent commented that

"Haem Arginate is poorly documented in the BNF".

The 23 people with porphyria attacks, but not hospitalised, treated themselves as follows:

Glucose	10 people
Pain killers`	3 people
Herbalist	1 person
Nothing	9 people

Delay in treatment for those hospitalised in the last 2 years

between arrival time in hospital, and receiving treatment.

less than an hour	6 people
4 hours	1 person
8 hours	1 person
On day admitted	3 people
48 hrs	1 person
4 days & 11 days	1 person (on two occasions)
Total	**13 people**

Generally treatment started promptly. There was sometimes a significant delay before haem arginate was given.

Satisfaction with treatment

	Hospitalised	Attack, not hospitalised	Other	**Total**
Satisfied	10	6	18	**34**
Unsatisfied	5	6	6	**17**
Totals	**15**	**12**	**24**	**51**

The majority were satisfied, but by no means all.

The greatest dissatisfaction was among those with attacks which were not severe enough to be hospitalised.

How could treatment be improved?

	not hospitalised	hospitalised	total
Better understanding	4	-	4
Better knowledge	12	4	16
Recognition of problem	1	2	3
Other	1	1	2
Total25	**7**	**25**	

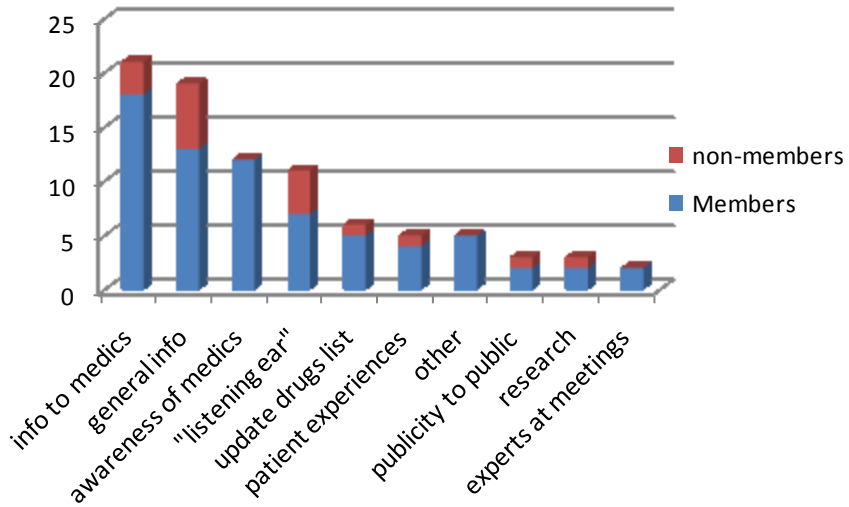
There needs to be better knowledge of porphyria among those treating patients.

What do you want from BPA?

	Members	non-members	All
information to medics	18	3	21
general information	13	6	19
awareness by medics	12	0	12
"listening ear"	7	4	11
up-to-date drugs list	5	1	6
patient experiences	4	1	5
publicity to public	2	1	3
research	2	1	3
experts at meetings	2	-	2
other 5	-	5	
Total suggestions	**70**	**17**	**87**
people making suggestions	**43**	**11**	**54**

We have tried to respond to these wishes – with our leaflets for patients and doctors.

Many of those saying they valued a listening ear, expressed satisfaction with the way we do that.



Acknowledgements

The BPA thanks all their members who kindly completed the questionnaire.
We are most grateful to Katherine von Gloss, who typed in the information from the forms into a database.

J. W. Chamberlayne
Chair, British Porphyria Association
March, 2009